

# Isolated ostial left main coronary artery stenosis causing ischemic cardiomyopathy in a child with bicuspid aortic valve: Role of echocardiography in diagnosis and follow-up

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## ABSTRACT

Bicuspid aortic valve (BAV) is the most common congenital cardiac defect, commonly associated with valve dysfunction and coarctation of aorta. Rarely, BAV can be associated with abnormalities of the coronary arteries, the most common of which are ectopic coronary origins. In this report, we present a case of a 2-year-old child with BAV, without coexisting supralvalvular aortic stenosis, who was found to have a left main coronary ostio-proximal stenosis, leading to ischemic cardiomyopathy and congestive heart failure.

**Keywords:** Bicuspid aortic valve, echocardiography, myocardial ischemia congestive heart failure

## INTRODUCTION

Bicuspid aortic valve (BAV) is a very common congenital heart defect, which can lead to a number of complications including aortic stenosis, aortic regurgitation, aortic dilation, and dissection.<sup>[1]</sup> Although not well understood, there appears to be a connection between BAV and coronary anomalies. Ostial stenosis of coronary arteries can develop as a result of certain congenital cardiac defects, namely, supralvalvular aortic stenosis (SVAS); this is an important complication because it is a risk factor for sudden death in patients with SVAS.<sup>[2,3]</sup> However, coronary ostial stenosis associated with a BAV remains exceedingly rare. This case demonstrates that coronary ostial stenosis can occur in association with a BAV and should be considered in cases of ventricular dysfunction.

## CASE REPORT

A term “infant” at birth had BAV with no stenosis or regurgitation, a circumflex right aortic arch with severe

coarctation due to severe hypoplasia of transverse arch, and isthmus with aberrant origin of the left subclavian artery from proximal descending aorta. The sinotubular (ST) junction measured around 7 mm with a Z score of +1. Due to concern about genetic syndromes, genetic testing was conducted, and the child was not syndromic. At 1 week, he underwent successful aortic arch reconstruction, coarctation repair, and vascular ring division. Subsequent follow-ups revealed aortic valve stenosis with a peak gradient of around 60 mmHg with a normal left ventricular (LV) ejection fraction (EF) of 60%.

At age 2 years, he presented with symptoms of respiratory infection, testing positive for rhinovirus/enterovirus. An echocardiogram revealed new onset LV dysfunction with an EF of 38%. Laboratory tests revealed brain natriuretic peptide of 4730 pg/mL. He was treated for congestive heart failure secondary

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to viral myocarditis and started on diuretics with inotropic support. During a routine echocardiogram 2 days after admission, abnormal flow pattern was noted across the left main coronary artery (LMCA) with color flow mapping and with Pulse Doppler imaging, a high-velocity flow pattern was visualized consistent with coronary artery ostial stenosis [Figures 1 and 2]. A cardiac angiogram confirmed the presence of severe coronary ostio-proximal LMCA stenosis [Figure 3]. After a subsequent surgical osteoplasty and aortic valvotomy, the patient had a LV ejection fraction (LVEF) of 48% with complete resolution of symptoms and residual aortic stenosis with a peak gradient of around 30 mmHg.

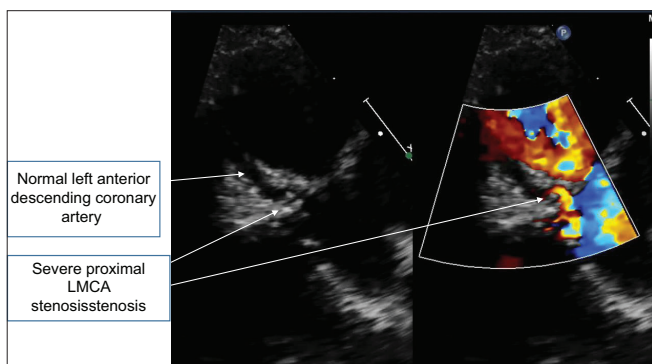
Seven months postoperative, he again presented with signs of diminished LV function and signs of congestive heart failure. Echocardiogram revealed significant LMCA ostial stenosis confirmed with cardiac catheterization. Cardiac magnetic resonance imaging (MRI) indicated diffuse subendocardial ischemic fibrosis with additional transmural scar in the inferoseptal LV wall [Figure 4]. During the subsequent surgical osteoplasty, there was ingrowth of scar tissue circumferentially and deep to the ostium toward the bifurcation of the LMCA. The coronary

was opened from the inside, and the scar tissue was removed. An implantable cardioverter-defibrillator was placed during surgery due to the extensive scar. His LVEF returned to normal postoperative, and AS has remained stable with a peak gradient of around 30 mmHg.

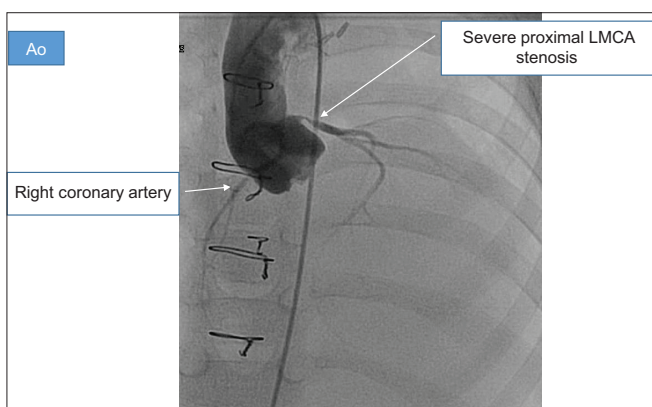
He is currently 5 year old and is well-being maintained on angiotensin-converting enzyme inhibitors and beta-blockers. The current LVEF is between 45% and 48% with no recurrence of ostial stenosis [Figure 5].

## DISCUSSION

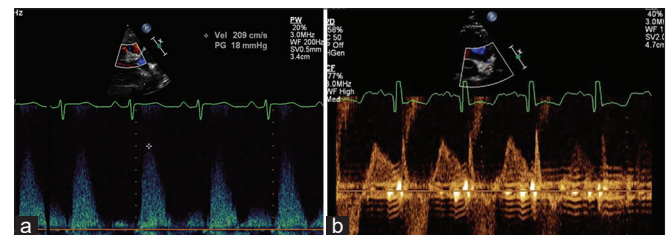
Coronary ostial stenosis in children can drastically reduce blood flow to a large part of the myocardium, leading to the development of ischemic cardiomyopathy. Ostial stenosis, such as that seen in SVAS, occurs in three subtypes; in Type I, the thickened media of the aorta extends to the ostium, resulting in a circumferential narrowing of the proximal LMCA. In Type II, the left coronary leaflet of the aortic valve fuses to the ST junction and blocks flow into the sinus of Valsalva and coronary artery. In Type III, there is diffuse narrowing of the LMCA to the bifurcation.<sup>[4]</sup> Each of these subtypes require different surgical strategies. The findings in our patient were typical for Type 1



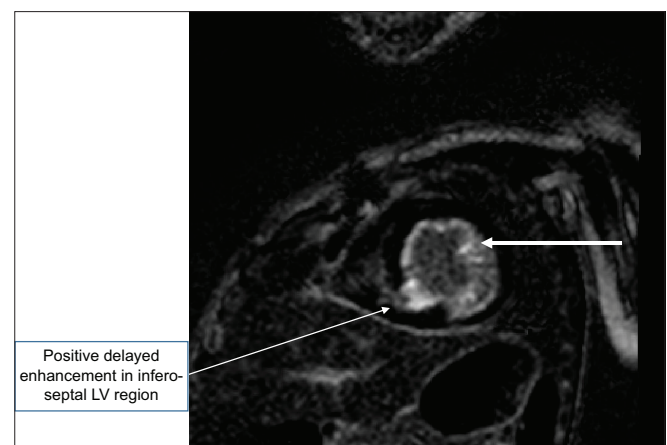
**Figure 1:** Parasternal long axis echocardiographic views show the left main coronary artery in 2-dimensional imaging and color Doppler mode to show severe proximal left main coronary artery stenosis



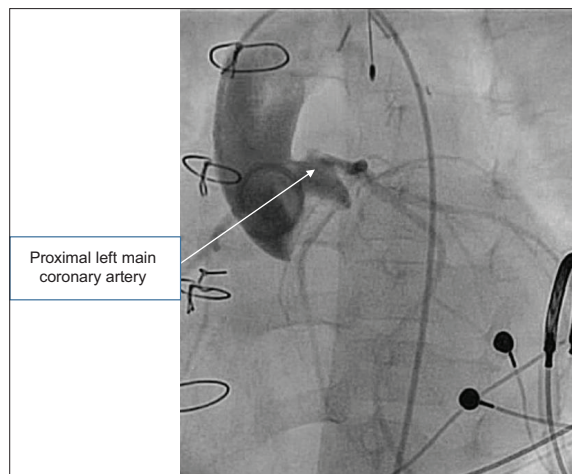
**Figure 3:** An angiogram in the anterior posterior projection (AP view) of the aortic root with normal right coronary artery and severe ostial stenosis and proximal left main coronary artery stenosis



**Figure 2:** (a) Pulse Doppler assessment of the proximal left main coronary artery with abnormal flow noted in both systole and diastole with peak systolic velocity of around 2 m/s suggestive of severe stenosis. (b) PW Doppler showing normal antegrade flow in the left main coronary artery during diastole above the baseline



**Figure 4:** Cardiac magnetic resonance imaging short axis views of delayed enhancement imaging shows scar (Fibrosis) in inferoseptal left ventricular region along with diffuse subendocardial fibrosis due to chronic ischemia (arrow)



**Figure 5: Aortic root angiogram in the AP projection shows a normal caliber of the left main coronary artery after second osteoplasty**

circumferential narrowing of the proximal LMCA. Type I is corrected by making an aortic incision that extends into the LMCA, then a patch is used to enlarge the aortic and ostial coronary stenosis. In Type II, the aortic leaflet is excised from the aortic wall, and a patch aortoplasty into the left coronary sinus corrects the aortic stenosis. Type III requires a coronary bypass to the left anterior descending and circumflex artery, then a patch aortoplasty that extends into the noncoronary sinus.<sup>[5]</sup>

In this case, the diagnosis of LMCA ostial stenosis was initially missed. Viral myocarditis was suspected until his symptoms did not improve with supportive treatment. It was not until the left coronary artery was investigated with echocardiography that a coronary abnormality was suspected as because of his symptoms. A cardiac catheterization confirmed the diagnosis and showed the extent of the narrowing and the caliber of the distal coronary arterial system.

Echocardiography can be an effective tool for assessing coronary involvement in patients presenting with unexplained ventricular dysfunction. Under normal conditions, the pulse wave Doppler in the LMCA shows a smaller systolic wave and a larger diastolic wave. The peak blood velocity in the LMCA should be  $24 \pm 6$  cm/s in diastole.<sup>[4]</sup> In ostial stenosis, there is an increased flow velocity noted during both systole and diastole. The peak systolic velocity in our patient was around 200 cm/s with the majority of flow noted during systole and less flow during diastole [Figure 2]. If abnormal flow is noted during echocardiography, findings should be confirmed with coronary angiography. Cardiac computed tomography and MRI may be useful, but coronary angiography is the most reliable imaging modality for assessing coronary arteries in children given the high spatial resolution and ability to assess the caliber of the distal vessels in case of potential bypass graft. Since cardiac MRI revealed nonviable myocardium and focal myocardial fibrosis, the

decision was made to implant an implantable cardioverter defibrillator (ICD) due to risk of sudden cardiac death in dilated cardiomyopathy with evidence of extensive fibrosis, especially in the septal regions.<sup>[6]</sup>

Restenosis of the coronary arteries can occur after angioplasty. Blood flow dynamics such as coexisting valve dysfunction that possibly contribute to ostial stenosis may still be present. As such, there is a need for long-term surveillance of coronary arteries. Periodic echocardiography is the best noninvasive imaging modality for this purpose. A routine assessment of not only the origin but also spectral Doppler assessment of flow can be a useful screening modality to screen for re-stenosis. Furthermore, serial pharmacological stress echocardiograms can be used in these patients to further assess coronary stenosis.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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